

» **Original Article** «

# Endovenous Radiofrequency Thermal Ablation and Ultrasound-Guided Foam Sclerotherapy in Treatment of Klippel-Trenaunay Syndrome

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**Introduction:** Klippel-Trenaunay syndrome is composed of port-wine stain, limb hypertrophy and varicose veins.

**Methods:** The two patients with Klippel-Trenaunay syndrome treated by endovenous radiofrequency thermal ablation and ultrasound-guided foam sclerotherapy of the abnormal veins was conducted.

**Results:** Radiofrequency thermal ablation resulted in successful occlusion of the incompetent anterior accessory great saphenous vein. Moreover, ultrasound-guided foam sclerotherapy showed complete occlusion of the residual veins. At 6 month follow-up, both patients markedly decreased leg symptoms including pain, cramping, limb swelling, and bulging of veins.

**Conclusion:** Radiofrequency thermal ablation combined with foam sclerotherapy is a minimally invasive procedure alternative to the standard invasive surgery and can be the option for saphenous ablation in Klippel-Trenaunay syndrome patients.

**Keywords:** RFA, foam sclerotherapy, Klippel-Trenaunay syndrome

## Introduction

Klippel-Trenaunay syndrome (KTS) is a mixed malformation that is characterized by abnormal development of veins, capillaries and lymphatics.<sup>1)</sup> The patient with KTS has the classical triad including limb hypertrophy, large varicose veins, and port wine stain (Fig. 1).<sup>1–3)</sup> The mainstay of therapy is conservative compression treatment including compressive bandaging and graduated compressive stockings.<sup>4)</sup> Duplex scan before surgical treatments on superficial veins must determine venous anatomy of deep vein

and confirm the patent deep venous drainage. The extensive surgical treatments including high ligation, venous stripping and phlebectomy have been the standard techniques for ablation of superficial veins and malformations in KTS.<sup>5–7)</sup> The minimally invasive endovenous thermal ablation including laser and radiofrequency have been reported in successful treatment of KTS.<sup>8)</sup> Ultrasound-guided foam sclerotherapy (UGFS) of abnormal large veins is also minimally-invasive technique and has been used in KTS.<sup>9)</sup>

In this study, the ablation of incompetence saphenous vein in KTS patients by endovenous radiofrequency (RFA) thermal ablation and UGFS was reported.

## Methods

A single center retrospective review of two KTS male patients treated with endovenous RFA of the anomalous KT veins and incompetence superficial veins was conducted. Preoperatively, the patients were

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**Fig. 1** Classical triad of Klippel-Trenaunay syndrome includes limb hypertrophy, large varicose veins, and port wine stain.

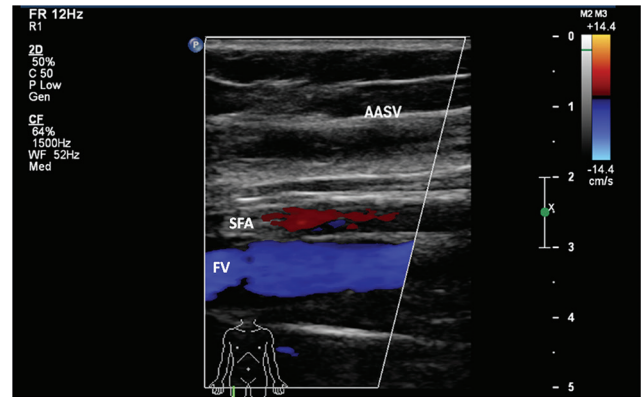
examined with color flow duplex ultrasound (GE LOGIQ9). The patients were also examined in a supine position with reverse Trendelenburg of 30 degree and standing position.

The venous malformations and lymphatic malformations including the truncular venous malformation (deep vein hypoplasia, marginal or lateral embryonic vein), extratruncular infiltrating venous malformation, truncular lymphatic malformation (primary lymphedema) and extratruncular infiltrating lymphatic malformation (lymphangioma) were evaluated in each patient.

Deep venous system was examined to demonstrate the patency of deep vein and exclude deep vein obstruction by compression maneuvers and both color and spectral Doppler analysis.<sup>4)</sup>

The anomalous KT veins and incompetent superficial veins were ablated with radiofrequency catheters (VNUS ClosureFAST; VNUS Medical Technologies, Inc., San Jose, California, USA). The RFA procedure utilized tumescent solution consisting of 450 cc of normal saline, 5 cc of 8.4% sodium bicarbonate and 45 cc of 1% lidocaine. Tumescent was then infused around the incompetence saphenous vein. Attempt was made to inject adequate tumescent to achieve a vein depth of 1 cm. The anomalous veins were cannulated with the RFA catheter. Temperature controlled treatment of 120 degree Celsius when using a 7 Fr catheter and treating 7 cm segments for 20 seconds was performed.

Concomitant 3% polidocanol in the form of foam sclerotherapy was injected into the residual varicose tributaries.<sup>9)</sup> The patients were asked to wear 20 to 30 mmHg graduated compression stockings. Additional



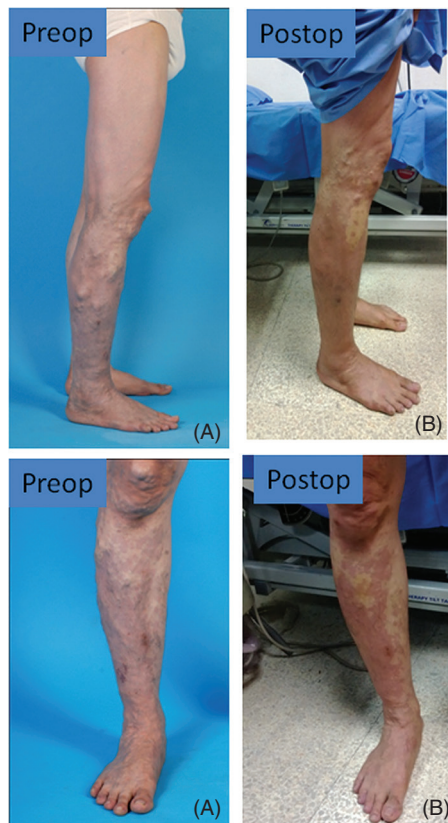
**Fig. 2** Duplex scan showed the patent of deep vein of the legs and anterior accessory great saphenous vein closed after radiofrequency thermal ablation.

follow-up was also consisted of venous duplex ultrasonography to evaluate the presence of deep system patency and continued ablation of the incompetent superficial veins. Ultrasounds were scheduled 3 days post ablation; 3 weeks post ablation, and then every 1 to 2 months for approximately 6 months. Multiple sessions of foam sclerotherapy was performed if the residual veins were still reopened.

This study protocol was approved by the Institutional Review Board.

## Results

Our retrospective study comprised of 2 male patients, aged 33 and 39 years. Both suffered from pain and swelling of the affected extremity. They had large varicosities and port wine stain and soft and bony tissue hypertrophy that developed in their childhood. Duplex ultrasonography of the lower extremities exhibited intact deep venous systems without any evidence of venous obstruction in both patients (Fig. 2). There was no reflux in GSV and deep venous system. Superficial reflux was demonstrated through anterior accessory great saphenous (AAGSV) in both patients. In the first patient, AAGSV connected to GSV at the knee level. In second patient, AAGSV connected to the large anomalous embryonic lateral veins of calf. AAGSV diameter ranged from 8 mm to 15 mm. The endovenous RFA was well tolerated by both patients. Duplex venous ultrasound after the procedures confirmed the thickening of the vessel walls with apparent complete occlusion of the AAGSV (Fig. 2). No deep vein thrombosis (DVT) was detected.



**Fig. 3** Post radiofrequency thermal ablation and multiple foam sclerotherapy showed decrease in varicosities and leg swelling. (A) before surgery, (B) post surgery.

Multiple ultrasound guided foam sclerotherapy of the large lateral anomalous varicose vein or residual varicose vein at calf was performed for 3 times in first case and 5 times for the second case.

The extratruncular infiltrating venous malformation, truncular lymphatic malformation and extratruncular infiltrating lymphatic malformation were not found in both patients.

The degree of varicose vein bulging, pain, and swelling improved following the procedure in both patients (Fig. 3). The first patient developed a small bruising at the puncture site that spontaneously disappeared in 7 days after the procedure. Neither thrombophlebitis nor skin burn was detected. Endovenous heat induced thrombosis (EHIT) or DVT did not occur during the 6 month follow-up period. The vessel is currently closed with the improvement of the patients' symptoms and appearance.

## Discussion

KTS is a mixed malformation with vein, capillary and lymphatic component. The management of KTS is composed of evaluation with diagnostic studies, treatment of clinical manifestations of KTS, prevention of vascular and orthopedic complications, and psychological support of the patient.<sup>8)</sup>

The KTS patients with hemorrhage, infection, acute venous thromboembolism, recurrent ulcer, pain, functional impairment, swelling due to chronic venous insufficiency and limb asymmetry require appropriate treatment. The management of KTS has been mostly conservative with compression therapy in the form of an elastic stocking or compression bandage. Compression therapy has been beneficial in treatment of lymphoedema and chronic venous insufficiency.<sup>8)</sup>

Surgery has been the classic therapy for vascular malformations. High ligation of the incompetent lateral marginal vein, venous stripping of superficial saphenous veins and ambulatory phlebectomy has been performed with using of a thigh tourniquet to decrease bleeding as the standard surgical treatment of KTS.

The endovenous RFA has been reported in successful treatment of KTS.<sup>4)</sup> The minimally invasive nature of the endovenous RFA procedure reduces bleeding and wound complications in KTS patients. RFA of the incompetent saphenous veins in KTS patients must be performed only when a patent deep venous system was identified by duplex ultrasound.<sup>4)</sup>

An intact deep venous system is important and must be established prior to performing RFA because some KTS patients do not have deep venous system.<sup>1,6)</sup> In such patients, the venous flow drains via only superficial veins that should be preserved to avoid serious consequences from severe venous hypertension.<sup>4)</sup>

The minor complication of RFA in our patient was a small bruising at the venous puncture site that was asymptomatic and disappeared spontaneously. There was no serious complication including skin burn, bleeding, EHIT, acute deep vein thrombosis and acute pulmonary embolism. Follow-up ultrasound showed complete ablation of the vessel.

Foam sclerotherapy with polidocanol has been used with success in KTS.<sup>8)</sup> We serially injected 3% Polidocanol in the form of foam sclerotherapy into the residual varicose veins to prevent technical failure from untreated collateral veins or feeding

varicosities that contributed to the recanalization of ablated segment.<sup>9)</sup>

Continued surveillance is required and adjunctive therapies such as foam sclerotherapy and additional thermal ablations may be necessary to treat the recanalized segments.<sup>4)</sup> The patient's symptoms were improved and remain stable at six month follow-up. So combined RFA and serial foam sclerotherapy was safe and effective in treatment of superficial reflux in KTS.

## Conclusions

RFA combined with foam sclerotherapy is a minimally invasive procedure alternative to the standard invasive surgery and can be the option for saphenous ablation in KTS patients. RFA combined with foam sclerotherapy appears to provide satisfactory cosmetic results and relief symptoms in short-term follow-up, a larger number of patients who are carefully selected over long-term follow-up should be further studied.

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## Disclosure Statement

All authors have no conflict of interest.

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